

# Clinical sustained uniform ventricular tachycardia in hypertrophic cardiomyopathy: association with left ventricular apical aneurysm

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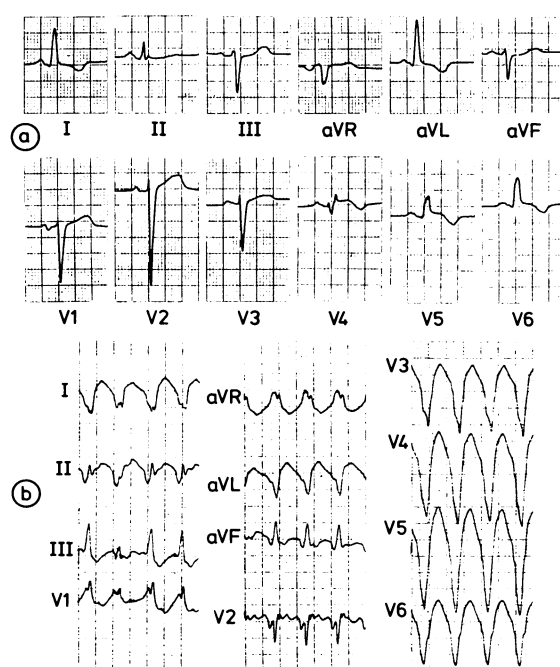
**SUMMARY** Of 51 patients with hypertrophic cardiomyopathy who had episodes of ventricular tachycardia detected during ambulatory electrocardiographic monitoring only two had clinical sustained uniform ventricular tachycardia that required medical treatment because of worsening symptoms. In both patients the arrhythmia was associated with the uncommon finding of an apical aneurysm with angiographically normal coronary arteries.

Ventricular tachycardia was detected during ambulatory electrocardiographic monitoring in 20-30% of unselected adults with hypertrophic cardiomyopathy.<sup>1,2</sup> The characteristics of these episodes was relatively homogeneous.<sup>3</sup> The ventricular tachycardia was slow, non-sustained, and asymptomatic, but it was a marker of subsequent sudden death.<sup>1-6</sup> Sustained ventricular tachycardia is rare and clinical characterisation of such patients has not been reported. We present two of 51 consecutive patients with ventricular tachycardia in whom episodes were rapid, sustained, and symptomatic; both patients also had a left ventricular apical aneurysm.

## Patients and methods

### CASE 1

Hypertrophic cardiomyopathy without a left ventricular gradient was diagnosed in a symptom free 40 year old man who presented in 1972 with an abnormal electrocardiogram (fig 1a). Angiography showed an apical left ventricular aneurysm with normal coronary arteries (fig 2). Two years later he presented with rapid palpitation and hypotension. Sustained

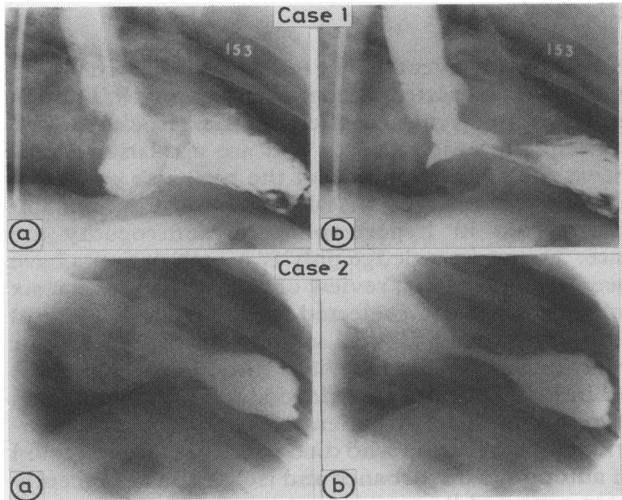


**Fig 1** (a) Electrocardiogram from patient 1 showing sinus rhythm with poor R wave progression in the right precordial leads and ST segment elevation in leads V4-V6. (b) Clinical sustained uniform ventricular tachycardia with right bundle branch block configuration. The axis is +160° and the QRS width 160 ms. A fusion complex (beat 2) is shown in the first panel.

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**Fig 2** Diastolic (a) and end systolic frames (b) from left ventricular angiograms (right anterior oblique view) from case 1 (upper) and case 2 (lower). Both patients show systolic mid-cavity elimination with increased residual contrast and paradoxical wall motion at the apex.

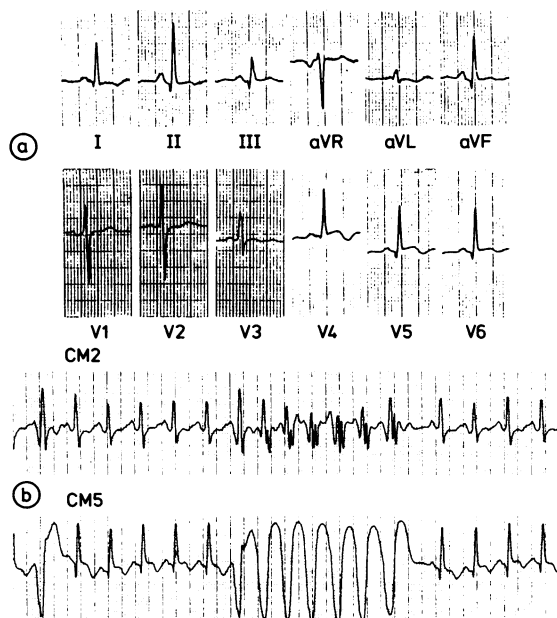
uniform ventricular tachycardia (180 beats/minute) required DC cardioversion. During a four week stay in hospital repeated episodes of sustained ventricular tachycardia with a consistent configuration (fig 1b) were not controlled with lignocaine, quinidine, or procainamide. An electrophysiological study was performed off medication in 1975. Sustained ventricular tachycardia with the same configuration could be repeatedly induced from the lateral border of the aneurysm and reproducibly terminated by two ventricular extrastimuli. Between 1975 and 1981 drug regimens that included disopyramide (2 g/day) plus propranolol (120 mg/day) and quinidine Durules (400 mg/day) plus verapamil (120–240 mg/day) were associated with gastrointestinal and neurological side effects, while dose reductions were associated with recurrence of the arrhythmia. Ventricular arrhythmias did not develop during maximal exercise tests. However, even during asymptomatic periods, episodes of non-sustained ventricular tachycardia were shown during electrocardiographic monitoring. In 1981 all other medications were stopped and amiodarone was started. A daily dose of 400 mg was required to maintain a minimum effective plasma concentration of amiodarone and desethylamiodarone (1.6 and 1.3 mg/l respectively).<sup>6</sup> For the past four years the patient has been symptom free without adverse effects or ventricular arrhythmia during electrocardiographic monitoring. Recent non-invasive evaluation confirmed the diagnosis and the abnormal anatomy. High gain, signal averaged electrocardiography performed by Simson's method<sup>7</sup> with high-pass filtering at 25 Hz showed a prolonged

filtered QRS duration of 150 ms (normal < 120 ms), a prolonged duration of terminal filtered QRS below 40  $\mu$ V of 41 ms (normal < 40 ms), and a reduced root mean square voltage in the last 40 ms of 18.1  $\mu$ V (normal > 25  $\mu$ V). Cross sectional echocardiography showed asymmetric septal hypertrophy with maximum wall thickness of 2.2 cm in the septum and left ventricular free wall at papillary muscle level. There was hyperdynamic contraction at the base of the left ventricle with mid-cavity elimination and an apical aneurysm which was best visualised from the apical two chamber view. There were no echocardiographic or Doppler features of a gradient but colour Doppler showed evidence of mild mitral regurgitation. Phase analysis of technetium-99m radionuclide ventriculography showed that the apex was 180° out of phase with the body of the left ventricle.

#### CASE 2

A 56 year old Indian man with familial hypertrophic cardiomyopathy and a history of exertional chest pain and syncope was referred after a syncopal episode associated with palpitation during which sustained uniform ventricular tachycardia was documented. The tachycardia was at a rate of 190 beats/minute with right bundle branch block configuration, an axis of  $-120^\circ$ , and a QRS width of 140 ms. Over a 40 year period he had experienced six other syncopal episodes that were not associated with chest pain or palpitation. Episodes of exertional chest pain began when he was 26, were often prolonged (1–2 hours), but were not associated with characteristic electrocardiographic changes of ischaemia or with increased plasma concentrations of myocardial enzymes. They were not consistently relieved by nitrates but treatment with verapamil was associated with fewer episodes (about one a month) than  $\beta$  blockers. An electrophysiological study was performed off medication in India. Single and double ventricular extrastimuli failed to induce sustained ventricular arrhythmias. Treatment with sotalol, verapamil, and verapamil plus procainamide failed to suppress episodes of non-sustained ventricular tachycardia during electrocardiographic monitoring.

Our investigations, which were performed after antiarrhythmic drugs were stopped, showed an abnormal electrocardiogram (fig 3a) and frequent ventricular extrasystoles (> 1000 daily) with daily episodes (1–5, median 3) of uniform non-sustained ventricular tachycardia at a rate of 160–180 beats/minute (fig 3b). Exercise testing was limited by fatigue and breathlessness at a maximal oxygen consumption of 26 ml/kg per minute and no arrhythmia or additional ST segment changes were seen. Cross sectional echocardiography showed asymmetric septal hypertrophy and a small left ventricular apical chamber that expanded during systole. The



**Fig 3** (a) *Electrocardiogram from case 2 showing sinus rhythm with a normal QRS axis and non-pathological Q waves in inferolateral leads. The pattern of ST segment abnormality and elevation in precordial leads V4-V6 is similar to that seen in case 1.* (b) *Continuous ambulatory electrocardiographic recording of an episode of non-sustained ventricular tachycardia that was similar in configuration to the sustained episode and to other non-sustained episodes.*

wall was thickest (2.4 cm) in the anterior and posterior septum and the free wall at the level of the tip of the mitral valve; there was incomplete systolic anterior motion of the mitral valve. The papillary muscles were greatly hypertrophied with systolic mid-cavity elimination that was associated with a mosaic pattern of turbulent flow on colour Doppler. At cardiac catheterisation there was no resting left ventricular outflow tract or mid-cavity gradient and the left ventricular end diastolic pressure was 26 mm Hg. Left ventricular angiography (fig 2) showed an akinetic area at the apex with dynamic contraction at the base; the coronary arteries were angiographically normal, apart from a separate origin of the left circumflex and systolic compression of septal perforator arteries. Radionuclide ventriculography with technetium-99m showed mid-cavity elimination with an apical aneurysm that on phase analysis was 180° out of phase with the remainder of the left ventricle. During the past year the patient was symptom free on amiodarone 400 mg daily with plasma concentrations of amiodarone and desethyl-amiodarone of 1.4 and 1.1 mg/l respectively. During 24 hour electrocardiographic monitoring there was a maximum of 240 uniform ventricular extrasystoles and no ventricular tachycardia.

## Discussion

Non-sustained ventricular tachycardia is common in adults with hypertrophic cardiomyopathy.<sup>1,2</sup> Though episodes are asymptomatic they are associated with subsequent sudden death and are a relatively sensitive and specific marker of the high risk patient.<sup>2-6</sup> Clinical episodes of sustained uniform ventricular tachycardia in hypertrophic cardiomyopathy are rare. To our knowledge only two such patients have been reported.<sup>8</sup> In a review of published reports, six other patients were selected for electrophysiological study because of clinical ventricular tachycardia or ventricular fibrillation, but no haemodynamic, echocardiographic, or additional arrhythmia data were available.<sup>9,10</sup> A patient with polymorphic ventricular tachycardia who died suddenly after a period of autonomic disturbance and ischaemia has recently been reported.<sup>11</sup>

Both of our patients with sustained uniform ventricular tachycardia also had a left ventricular aneurysm with angiographically normal coronary arteries. The apical aneurysms were shown angiographically and confirmed by echocardiographic and radionuclide studies. We did not see this configuration in the 49 patients with non-sustained ventricular tachycardia or in over 200 patients with hypertrophic cardiomyopathy without ventricular tachycardia who were similarly evaluated. Apical aneurysm in hypertrophic cardiomyopathy has been reported but is rare and is usually associated with normal coronary arteries.<sup>12-14</sup> A recent report suggested that the presence of mid-ventricular obstruction may predispose to wall motion abnormalities in the distal left ventricle.<sup>13</sup> When our patients were first examined there was no evidence of a left ventricular gradient; however, mid-ventricular obstruction earlier in their course cannot be excluded. Alternatively the apical myocardial changes may have been caused by coronary artery spasm with myocardial infarction.<sup>14</sup>

The electrocardiograms in our patients showed diagnostic clues with precordial ST segment elevation in the absence of abnormal Q waves. This pattern was reported by Gordon *et al* in three patients with apical aneurysm.<sup>15</sup> Similar electrocardiographic changes were not seen in a review of electrocardiograms from 100 consecutive patients with hypertrophic cardiomyopathy who did not have left ventricular aneurysm.<sup>16</sup> We agree with Gordon's suggestion that these electrocardiographic features may be a marker of ventricular aneurysm.<sup>15</sup> The patient in whom high gain, signal averaged electrocardiography was performed (case 1) had late potentials. The association between the presence of late potentials and a propensity to ventricular tachycardia, especially in patients with left ventricular aneurysm, has been well established,<sup>17</sup> though the relevance of such a finding in patients with hypertrophic cardio-

myopathy has not yet been systematically investigated.

Sustained uniform ventricular tachycardia is also uncommon during programmed electrical stimulation studies for the evaluation of clinical arrhythmias or identification of high risk patients with hypertrophic cardiomyopathy.<sup>8-10 18-24</sup> More commonly multiform ventricular tachycardia or ventricular fibrillation or both is induced. The clinical significance of these induced arrhythmias is controversial.<sup>8-10 19-24</sup> Recent studies have emphasised that to enhance the specificity of the results less vigorous protocols should be used.<sup>8 20 24</sup>

The prognostic significance of clinical sustained uniform ventricular tachycardia in hypertrophic cardiomyopathy, whether or not it is related to the presence of apical aneurysm, remains to be established. Both patients are symptom free and without recurrence of arrhythmia during long term treatment with amiodarone. Should it be necessary, surgical aneurysmectomy remains an option.

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